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# LQTS: Gender Differences and Mother-Son vs. Mother-Daughter

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# LQTS: GENDER DIFFERENCES AND MOTHER-SON VS. MOTHER-DAUGHTER PERCEPTIONS OF QUALITY OF LIFE

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**Dissertation Approval**

This is to certify that the thesis presented to us by Thea Gallagher on the 29th day of April, 2014, in partial fulfillment of the requirements for the degree of Doctor of Psychology, has been examined and is acceptable in both scholarship and literary quality.

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## Abstract

Long QT syndrome is a chronic disorder, and one of the most common genetic arrhythmia syndromes that can cause unexpected cardiac arrest and death in individuals. Treatment options include medications, use of implantable cardioverter defibrillators (ICD) and restricting activities of daily life. Minimal research exists exploring the specific psychosocial aspects of the illness and/or the impact on the quality of life of individuals and families coping with the management and diagnosis of this illness. Furthermore, little research has attempted to understand the role of gender differences and mother-child perceptions among individuals with Long QT syndrome. The current study used data from a larger study in an effort to specifically examine overall gender differences in QOL, as well as perception differences between mother-son and mother-daughter perceptions of QOL. The study predicted that mothers and sons would have less matched perception with regard to QOL domains, while mothers and daughters would be more aligned. The study also predicted that female children would have a higher self-reported QOL than male children. Results found that there was greater cross-informant variance for sons and mothers on the treatment anxiety subscale but not statistically significant findings for overall cross-informant variance. There were no statistically significant findings for gender differences overall, but results suggested that women were indicating a lower QOL than males. Results also found that emotional functioning was identified as lower than the other subscales, and lower than in healthy individuals, indicating the emotional needs of those with Long QT syndrome may be compromised. Further research addressing the psychosocial needs of children and families with Long QT syndrome is necessary.

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# Chapter 1

## Introduction

### Statement of the Problem

Chronic illness affects a significant number of children and adolescents in the United States (Ingerski, Shaw, Gray, & Janicke, 2010). Research indicates that up to 19.3 percent of children in the United States have specific health care needs (Ingerski et al., 2010). This data does not fully explain the broad psychosoial burden that families of chronically ill children and youth bear. Research suggests that there are significant negative psychosocial symptoms that chronically ill children and their parents experience in coping with a particular pediatric disease (Ingerski et al., 2010). Chronic illness has been defined as a disease with an extended course in which individuals can maintain normal lifestyles, or as a disease that can progress toward a negative and sometimes fatal end (Kanney, Hamrin, & Grey, 2003). Families affected by chronic illness must deal with multiple obstacles and repercussions that accompany the management of a chronic condition (Crespo, Carona, Silva, Canavarro, & Dattilio, 2011).

Long QT Syndrome is a chronic disorder with symptoms ranging from none to sudden death (Giuffre, Gupta, Crawford, & Leung, 2008). The syndrome is defined as a constellation of repolarization disorders, in which the corrected QT interval is prolonged (Farnsworth, Fosyth, Haglund et al., 2006). This chronic condition can lead to life-threatening heart arrhythmias. LQTS may also lead to sudden death in children and young adults who have not been diagnosed and are not aware that they have the

syndrome (Farnsworth et al., 2006). Data suggest that individuals and families affected by LQTS experience a variety of psychosocial challenges (Giuffre et al., 2008).

Since chronic illness is a persistent threat (particularly LQTS, with the unpredictability of cardiac arrest and death), it can dominate the family system, structure, and function, and therefore other developmental needs of the family may be neglected (Cohen, 1999). Children with continuing health conditions are at greater risk of mental health problems, and the odds of a psychiatric disorder in a child with chronic illness are about 2.2 to 1 (Cohen, 1999). This fact alone sheds light on one of the many issues that have to be faced in regard to the physical and mental health of chronically ill children and their families.

**Purpose of the Study**

The purpose of this study is to evaluate the relationship between between male and female children with LQTS and their mothers. The study aims to investigate quality of life as perceived by male and female children with LQTS and the parent's perception of the child's quality of life. This investigation will attempt to determine if there are differences in perceptions of quality of life between mother-son and mother-daughter dyads, and between male and female children. The study will compare the mother-reported and child-reported quality of life of children with LQTS, and will compare male children to female children. The connections between mothers and their children and male and female children will be assessed through two measures, which examine the mother's perception of her child's coping and the child's perception of their own coping. Research has found that children with cardiovascular disease have lower perceived quality of life in comparison to healthy children (Uzark, K., VonBargen-Mazza, P., &

Messiter, E, 1989). If their primary caregiver does not perceive a child's quality of life accurately, the caregiver may be less likely to understand the severity of the psychosocial issues, which could have a negative impact on the family functioning and overall quality of life of both the children and the caregivers. A better understanding of mother-son and mother-daughter perception of quality of life and coping, as well as gender differences, will increase the knowledge base on this topic. With an increased knowledge base, more effective efforts in skills training, psycho-education, and family therapy can be facilitated for this population.

# Chapter 2

## Literature Review

### Chronic illness.

Data suggest that up to 19.3 percent of children in the United States have special health care needs (Ingerski, Shaw, Gray, & Janicke, 2010). The myriad practical concerns and the overall burden on families of chronically ill individuals have been under-represented in the literature. Research suggests that children and their caregivers experience significant psychosocial symptoms while trying to cope with a particular pediatric disease (Ingerski et al., 2010). The aforementioned psychosocial difficulties may negatively affect treatment adherence, but there is little research explaining child and family functioning in a cohesive framework for chronically ill populations. Some research has suggested that the diagnosis of a chronic illness can be a traumatic event that may lead to PTSD (Ingerski et al., 2010). Researchers across a number of chronic illnesses have found the parents and children report traumatic stress symptoms in response to the diagnosis of the child (Ingerski et al., 2010). Data suggest that individuals diagnosed with chronic illness perceive themselves as having a lower quality of life, indicating that they have less hope of their situation improving (Panides & Ziller, 1981).

### LQTS

When a healthy young individual dies suddenly and unexpectedly, it is a devastating event with severe consequences for caregivers, family, and community. Long QT syndrome is one of the most common genetic arrhythmia syndromes that can cause unexpected cardiac arrest and death in a young person. Long QT syndrome (LQTS) is a

congenital disorder, which may produce life-threatening arrhythmias, syncope, aborted cardiac arrest, and sudden cardiac death, which is usually a result of ventricular fibrillation (Anderson, Oyen, Bjorvatn, & Gjengedal, 2008). LQTS is characterized by abnormal QT interval prolongation on an ECG (Roden, 2008). The disease is most commonly inherited in an autosomally dominant fashion (Anderson et al., 2008).

There are three main phenotypes for LQTS. In LQT1 patients, most life-threatening situations occur either during exercise or during emotional arousal (Anderson et al., 2008). Sudden or surprising noises, such as a fire alarm, can trigger a cardiac event in an LQT2 patient (Anderson et al., 2008). In LQT2 and 3 patients, these cardiac events can happen during sleep or rest (Modell & Lehmann, 2006). LQTS is predicted to affect about one in 2,000 to one in 5,000 individuals. The ratio could potentially be as high as 1 in 2,00 (Schwartz, P. J., Priori, S. G., Spazzolini, C., Moss, A. J., Vincent, M., Napolitano, C., 2001;Tester et al., 2006)

If untreated, 6 to 13% of individuals with LQTS succumb to cardiac arrest or sudden cardiac death before the age of 40 (Anderson et al., 2008). In 12% of those cases, cardiac arrest or sudden cardiac death was the first manifestation of the disease (Anderson et al., 2008). Treatment options are medications, specifically beta-blockers, or use of implantable cardioverter defibrillators (ICD) for those who have repeated syncopes, or who have needed resuscitation after cardiac arrests (Hendriks, et al., 2005). Restricting activities of daily life (ADL) such as avoiding mental stress, physical exhaustion caused by work or sports, and avoiding loud noises, is also generally advised (Hendriks et al., 2005)

Although about 90% of LQTS cases are inherited and all are related to genetics, about 10% are sporadic, or manifested through use of medications, such as antibiotics, antidepressants, antihistamines, diuretics, antiarrhythmic medications, cholesterol-lowering drugs, diabetes medications, as well as some antifungal and antipsychotic drugs that lengthen the QT interval in otherwise healthy individuals (qtdrugs.org; MayoClinic.com). LQTS also has a significant effect on the caregivers and siblings of a diagnosed individual (ten Kroode, et al., 2000). It has been found that family members who do not have LQTS are just as troubled as those who suffer from LQTS. These individuals remain anxious and worried, as they remained connected and concerned about their affected relatives (ten Kroode et al., 2000). It has also been found in some cases, that parents are more focused on the wellbeing of their children than on their own health (Hendriks et al., 2005). In the few studies that have addressed psychosocial concerns in LQTS patients and their families, high levels of distress have been found, although there is very little detailed research on this population and the challenges they face (Anderson et al., 2007).

**Risks.**

Parents and children affected by a chronic health condition are at risk for decreased psychological functioning, wellbeing and quality of life (Crespo, Carona, Silva, Canavarro, & Dattilio (2011). In regard to psychological adjustment, children with a chronic condition are more likely to have anxiety disorders, behavior problems, and low self- esteem (Crespo et al., 2011). Parents of children with a chronic health condition are also at risk for depression, anxiety and emotional distress (Crespo et al., 2011). Children with LQTS in specific have been found to score high on internalizing and externalizing

behaviors, which further suggests that these children are unable to discuss feelings of anxiety and subsequently cannot cope with a discussion of those feelings in an open forum (Giuffre, Gupta, Crawford, & Leung, 2008).

Families affected by congenital heart disease are often exposed to in-depth discussions regarding risks connected to the progression of the disease, risks of diagnostic interventions, and intraoperative and postoperative complications (Giuffre et al., 2008). For the LQTS population, these discussions address the risk of sudden death and use of measures to prevent its occurrence, such as an Automated External Defibrillator (AED) (Giuffre et al., 2008). The manifestation of LQTS is so broad (ranging from no symptoms to sudden death) that it is expected that patients with LQTS and their parents would experience significant anxiety and fears (Giuffre et al., 2008).

There are few studies that look at the psychosocial aspects of LQTS, but the preliminary studies have indicated that families affected by cardiac heart arrhythmias have reported high levels of distress (Giuffre, Gupta, Crawford, & Leung, 2008). In light of the fact that specific biopsychosocial research is lacking for this population, it is helpful to look at the broad spectrum of chronic illness factors associated with individual, family, and caregiver quality of life.

## Strain on Family

### Parents.

Caregivers are greatly impacted by chronic illness, especially those of their child and/or children. Given the genetic nature of LQTS, there is a significant probability that more than one child will be diagnosed with the syndrome, 50% for each child, magnifying the care needed. Research has indicated that parents of children with a

chronic illness perceive daily life as more stressful than parents of healthy children (Crespo et al., 2011). The responsibilities placed on the caregiver include the care of the child's physical, medical, and emotional needs. The caregiver also is responsible for helping the child cope with the physical and emotional demands of their condition (Crespo et al., 2011).

Parents as caregivers of chronically ill children are at increased risk for stresses and strains, vulnerability to depression, and marital discord (Cohen, 1999). One of the overarching difficulties of living with a child with a chronic illness is uncertainty about the recurrence of the disease even after treatment, and in the case of LQTS, there is always the probability of an unforeseen cardiac event (Sloper, 2000). Recurrent threat of a cardiac event, or in some cases, misfiring of the implantable cardioverter-defibrillator, (ICD) can intensify negative effects of stress for parents (Cohen, 1999). It was found that the greatest fear for parents of children with LQTS is that something detrimental might happen to their children, such as a cardiac event (Anderson et al., 2008). These are important psychosocial demands to examine with caregivers of children with LQTS.

Research suggests that parents struggle to judge their child's functioning impartially, because their own experience of traumatic stress may influence how they perceive their child's overall adjustment and coping (Ingerski et al., 2010). There are data implying that children and families affected by LQTS may experience some traumatic stress symptoms (Ingerski et al., 2010). These PTSD symptoms may have a pervasive impact on the overall quality of life of children and families affected by LQTS. The present study will be analyzing mothers of children of LQTS, but the information about

both parents helps to provide a greater understanding of gender differences in coping at the adult level.

There is limited research about the mental health of parents of chronically ill children, and even less research about parents of LQTS-diagnosed children (Cohen, 1999). Parent-child communication in the LQTS population has not been studied. Gender differences in parental coping have been studied through historic interactions and research methodology (Cohen, 1999). In that research, maternal reports tend to be considered as "total family functioning," because mothers are traditionally the main caregivers and the participant most easily available to researchers (Cohen, 1999). Mothers are interviewed more frequently by researches; therefore, the information surrounding the issue of parental psychosocial concerns may not be representative of both males and females. Increasing evidence has shown that childhood illness has significant effects on fathers and the total family system and should be an area for increasing research (Drotar, 1992).

**Parents/Gender Differences.**

Differences have emerged between mothers and fathers of children with chronic illness: mothers have more involvement in treatment and more interaction with the health provider; they provide more caregiving at home; and the chronic health problems of the child correlated with higher distress in mothers than in fathers (Quittner, Espelage, Opipari, Carter, Eid, & Eigen, 1998). Research on fathers' coping has shown greater stress in relation to finances and emotional attachment to the child (Quittner et al., 1998). Mother/father coping differences imply that there are gender differences that may influence perceived stress and coping patterns (Cohen, 1999). One study found that

mothers and fathers of children with diabetes have unique vulnerabilities and different ways of coping with chronic illness (Dashiff, 1993) The findings indicate that mothers had a tendency to use strategies to control the illness and its treatment plan, but fathers were more likely to use distraction or distancing to cope with illness stressors (Dashiff, 1993). This information is important for the evaluation of mother's perception of child coping, because mothers may have different ways of coping that may consequently influence perception (Sloper, 2000).

**Family Cohesion**

It has been found that family cohesion is an important resource for mothers and fathers; higher levels are related to greater adjustment of parents and children (Mastroyannopoulou, Stallard, Lewis, & Lenton 1997). Families with strong relationships can even become closer through illness, in contrast to families with weaker relationships that may assume that the illness caused the problems (Mastroyannopoulou et al., 1997)., Family cohesion also can be degraded by stresses, such as role conflicts over caregiving and subsequent family-functioning issues that are not addressed by caregivers (Cohen, 1999). Some of the tensions that can threaten parental cohesion are financial strains, loss of family privacy and freedom, the personal stress of caring for a sick child, worries about the future, the consequences of constant pressure, and problems with health-care staff and insurers (Cohen, 1999). Other threats are extended periods of existential loss, including profound guilt, regret over the "normal" life they and their children might have had, and in some cases, anguish over causing their child's suffering through genetic transmission (Cohen, 1999).

As family systems theory posits, relationships between family process and illness are of a reciprocal nature (Cohen, 1999). Family processes may have negative or positive interaction effects (Cohen, 1999). It has been found that assessing quality of life, reciprocal interactions between family factors, illness, and patient biopyschosocial outcomes can inform assessment and enhance supportive interventions (Cohen, 1999). Assessment through this lens also addresses the potential for negative impacts by these factors on the illness (Cohen, 1999).

## Children/ Gender differences

### Parents/Children and Communication

Since there is much research to support the idea that family systems are greatly affected by chronic illness, it is important to examine certain risk factors that could be exacerbated by an LQTS diagnosis. There is a wealth of research establishing that conflict in families is correlated with negative outcomes for children (e.g., low self-esteem, depression, drug abuse, or delinquency) (Beaumont, Vasconcelos, & Ruggeri, 2001). Communication difficulties were found to contribute to conflict and the subsequent negative outcomes in families. Successful conversations were found to occur more regularly between individuals who share the same communication styles (Beaumont et al., 2001). Individuals who are similar in style of communication share expectations for the pace of taking turns or simultaneously speaking, but speakers with dissimilar styles tend to have different expectations and therefore more likely to interrupt and talk over each other (Beaumont et al., 2001). It has been found that mothers and sons use dissimilar styles of communication more than mothers and daughters, although there is some variability across developmental stages (Beaumont et al., 2001). Communication between

individuals is enhanced when a common base of experience can be built upon. It also has been found that communication is enhanced when personalities are similar. This information has implications for families communicating about topics relating to chronic illness, which can dominate the family system.

Research has found that parental perceptions of children's abilities in a number of domains are positively related to a child's performance academically and socially (Flannagan & Baker-Ward, 1996). It has been established that parent-child discourse about children's experiences influences the development of children's competence and self-understanding (Flannagan & Baker-Ward, 1996). Parental feedback has been found to help children review and receive information that impacts the child's impression of those experiences.

Parental monitoring is another aspect of chronic illness management that is important (Hafetz & Miller, 2010). Parental monitoring examines the parenting behaviors related to the parent's involvement in and oversight of the child's activities (Hafetz & Miller, 2010). If there are problems in parent-child communication, parental monitoring is negatively impacted. Parents help children adhere to treatment regimens. If parents do not communicate with their children about details of treatment adherence, negative health outcomes may result (Hafetz & Miller, 2010). In the case of LQTS, parents have to oversee a treatment regimen and balance that with other child and family responsibilities and activities (Hafetz & Miller, 2010). It is therefore important to understand the nature of dyadic communication and interaction in order to increase treatment adherence and overall health and QOL.

**Mother/Child Interaction**

Mothers spend more time talking to their daughters about emotions, and sons have been found to be more withdrawn in conversations with their mothers than daughters (Beaumont et al., 2001). Positive emotional attachments between parents and children are correlated with increased cohesion and adaptability in their interactions, and the child's individuality and connectedness are enhanced (McKinney & Renk, 2007). Data suggest that parents may employ different parenting behaviors based on the gender of their children and adolescents (McKinney & Renk, 2007). Mothers tend to spend more time being accessible to their children and take more responsibility for their children's activities than fathers. In chronic illness populations, the mother is often the caregiver, and subsequently spends more time with the diagnosed child than the father. It therefore appears that boys are at greater risk for insufficient communication and subsequent negative outcomes in relationships.

Parenting plays an important role in the adjustment of healthy children. Research indicates that parenting plays an even more crucial role in the life of a child with chronic illness (Hegarty, MacDonald, Watter, & Wilson, 2008). In studies that have examined late adolescents, it was found that mothers' parenting was a significant predictor of female late adolescents' adjustment (McKinney & Renk, 2007). Females tend to view their mothers as providing a more mutually supportive relationship compared to their males counterparts (McKinney & Renk, 2007). Subsequently, it is assumed that mothers' parenting is an influential predictor of female adjustment in late adolescence, even in the context of such characteristics of the family environment as conflict and the management of chronic illness (McKinney & Renk).

Research indicated that a positive family environment is beneficial to the adaptation to children with a chronic condition (Crespo et al., 2011). Parents' positive perception of their child's QOL positively informs the child's perception of their own QOL (Crespo et al., 2011).

**Pain Perception.**

The perception and expression of pain can be used as a guide to understanding management of chronic illness symptoms. With LQTS, the pain experience can range from dormant symptoms to cardiac arrest. The uncertainty of the LQTS pain symptoms has been found to have negative effects on individuals with the illness (Janney, 2011).

One aspect of chronic illness involves the connection of pain perception to gender differences. Even though children with LQTS do not experience physical pain regularly, the research provides insight into differences in gender coping and communication. It has been found that boys report less pain perception than girls (Vierhaus, Lohaus, & Schmitz, 2011). Gender differences in reports of pain may be influenced by gender-role expectations (Vierhaus et al., 2011). Biological factors may influence gender differences, but cognitive and psychosocial factors have also been found to play a role. It has been found that rigid gender-role expectations influence the likelihood of a boy to tolerate pain and report less pain experience (Vierhaus, 2011). Social pressure to conform to gender roles increases as children move through puberty, because it is rarely addressed and corrected in childhood (Hafetz & Miller, 2010). It has been well established in research findings that girls report more physical and psychological symptoms than boys (Vierhaus, 2011). Men also tend to cope with pain by using behavioral distraction in contrast to

females (Hechler et al., 2010). It has also been discovered that females tend to perceive less ‘”treatment burden” than males (Hegarty et al., 2008).

It has been further suggested that gender differences in coping with pain experiences may be observed in differing parental behavioral responses to a child with chronic illness (Hechler et al., 2010). Mothers have been found to report more discouraging responses about boys than girls, such as not taking their pain seriously or being hesitant to believe them (Hechler et al., 2010). It has also been suggested that mothers may be less likely to take their sons’ pain seriously, because they expect their sons to endure pain and adjust their pain behavior accordingly (Hechler et al., 2010). The discrepancy between mothers’ and sons’ perceptions and expressions of pain may affect the communication between mothers and sons about these issues, which could have a negative effect on the child’s overall quality of life.

**Gender Communication Differences Among Children**

A number of studies indicated that girls seek more social support and use more emotion-focused strategies than boys (Calvete, Camara, Estevez, & Villardon, 2011). Data also suggest that boys use more avoidant coping, in which less social support is sought (Calvete, et al., 2011). Research indicated that social support is a strategy utilized more consistently by girls than boys (Piko, 2001). It has been found that passive and risky coping factors play a negative role in psychosocial health, but problem analyzing and support seeking play a positive role (Piko, 2001). Participation was also found to be a positive correlate with coping, which has implications for the LQTS population, whose exercise is typically restricted. This information has further implications for boys with

LQTS; exercise may be their main source of coping behavior, which would be affected by the diagnosis.

Boys have been found to have less trust and more reluctance to turn to others as a source of support (Piko, 2001). Boys are more inclined to manage issues independently, which may negatively affect their psychosocial health, especially when managing the aforementioned issues associated with an LQTS diagnosis (Piko, 2001). Data suggest that girls use social support more frequently, yet boys may actually derive greater benefit from this coping strategy (Piko. 2011). In regard to emotional regulation strategies, girls use less distraction/recreation and more aggression (e. g., verbal expression, agitation, expressing frustration) than their male counterparts (Eschenbeck, Kohlmann, Lohaus, 2007). Characteristics of the stressful situation influence gender differences in coping in children and adolescents (Eschenbeck, 2007). In a stressful situation such as LQTS, it can be posited that these behaviors have more impact and may be associated with greater vulnerability on the part of the child.

**Quality of life affected by coping**

Coping for this population has been defined as “parent's perception of how he or she manages family life with a child with a chronic illness’”(McCubbin, McCubbin, Patterson, Cauble, Wilson, & Warwick, 1983). How parents respond to the stress and anxiety associated with the illness of their child is important to understanding how they perceive their overall management of their family and of the sick child. Parental coping, as previously explored in the research, contributed to positive family functioning, quality health care from parents at home, and positive improvements in the health of the child (McCubbin et al., 1983).

**Avoidant coping, Emotion-focused coping, and Problem-focused coping.**

Understanding what factors place individuals at more or less risk for low quality of life after being diagnosed with LQTS is important for prevention and treatment efforts (Hassan, et al., 2011). From a broad perspective, coping is defined as thoughts and behaviors used by an individual to minimize distress and manage its emotional impact (Folkman & Lazarus, as cited in Hassan et al., 2011). One theory of coping presented in the literature is the triadic model of coping, which includes emotion-focused, avoidance-focused, and problem-focused coping styles (Hassan et al., 2011). The triadic model provides insight into gender differences in coping styles and identifies different aspects of coping linked to coping strengths and weaknesses in males and females.

Emotion-focused coping is when an individual has an awareness of their emotional distress, and they are able to vent or release those feelings (Hassan, et al., 2011). Rumination is a component of emotion-focused coping, in which individuals engage in repetitive thoughts focusing on symptoms of distress and causes and outcomes of such symptoms (Hassan, 2011). People who engage in emotion-focused coping can fixate on the problems and distressing symptoms instead of looking for ways to problem solve (Hassan, 2011). Some studies suggest that women utilize emotion-focused coping more than men (Hassan, 2011).

Avoidant coping strategies involve an individual's avoidance of identifying and managing stressful circumstances in his or her life (Hassan, 2011). These strategies can contribute to impulsive behaviors and can be characterized by denial, psychological dissociation, and behavioral disengagement (Hassan, 2011). Research indicates that men

may be more likely to employ avoidant coping strategies than women (Hassan, 2011). Avoidant coping has also been linked to higher rates of depression (Hassan, 2011).

Problem-focused coping involves an active engagement in attempting to change or manage a stressful situation. This coping strategy includes positive reinterpretations, positive cognitive reframing, planning, and seeking emotional and social support from others (Hassan, 2011). Higher problem-focused coping strategies have been linked to lower levels of depressed mood and subsequent higher QOL (Hassan, 2011). Lower QOL appears to be informed by reliance on employing problem-focused coping strategies.

**Quality of life**

Quality of life is defined by the World Health Organization (WHO) as "individuals' perceptions of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns" (1994). It has been found across age groups that children with cardiovascular disease perceive themselves to have a lower quality of life than healthy individuals (Uzark et al., 1989). QOL is degraded by the many and various stressors associated with LQTS and with caring for a child with LQTS. Health-related quality of life (HRQOL) has been identified as an important health outcome in clinical trials, in clinical improvement strategies, in population-based health assessment, and among consumers of health-care services (Varni, Seid, Knight, Uzark, & Szer, 2001). HRQOL includes physical-functioning dimensions, as well as psychosocial health dimensions of emotional, social, and role functioning (Varni et al., 2001). This is important; psychosocial health has been identified as the "new hidden morbidity" in pediatric healthcare, because it has been historically underidentified in pediatric practice (Varni et al., 2001). Pediatric HRQOL

measurement instruments should include a range of cognitive developmental stages, as well as both child self-report and parent-proxy report in an effort to understand potential differences in perspectives (Varni et al., 2001). Disagreement between self-report and proxy-report, also called cross-informant variance, has been consistently identified in child/adolescent and parent reports in the HRQOL assessment of children with chronic health conditions (Varni et al., 2001). Disagreement is higher about internalizing problems, such as depression and pain, and lower for externalizing problems (Varni et al., 2001). Considering that HRQOL comes from an individual's perceptions, the issue of cross-informant variance reveals the importance of reliable and valid pediatric HRQOL measurement for a broad age range (Varni, et al., 2001). In an endeavor to understand the differential impact of LQTS on the child and family, it is crucial to identify any discrepancies between parent and child perspectives on the child's QOL (Hegarty, MacDonald, Watter, & Wilson, 2008).

**Importance of Research and Future Research**

Understanding the adjustment of parents of LQTS patients is important in assessing and treating this specific population. Gender differences between parents and children can enhance awareness of the processes influencing the heterogeneity of responses (Sloper, 2000). More research should explore the unique experiences of caregivers and children affected by chronic illness, particularly in regard to coping and QOL. Coping differences between genders should be examined more fully, so that psychologists in medical health settings and those who interact with these clients in other ways are well equipped to meet the individual needs of these parents and children, and to improve overall family functioning in this vulnerable population. This information could

greatly help parents and children who may need additional support in any of these individual areas. With the LQTS diagnosis, parents and children must deal with the continuous threat of this life-endangering condition and the psychological consequences associated with it (Hendriks, 2005). Unlike some other serious childhood illnesses, LQTS does not go into remission, and the first event may cause death. Helping these parents and children adjust in the long-term is of great importance, for they will potentially have to manage this threat through their entire lives.

The present study is part of a larger, ongoing study that aims to examine QOL in children with LQTS and their parental caregivers. The study aims at developing a model explaining anxiety, depression, coping, and QOL in children with LQTS and their parents.

Understanding the unique coping challenges and QOL differences will be important for research connected to gender differences in children and to the needs of fathers and mothers dealing with the chronic illness of their child. Screening can help clinicians create appropriate interventions for families in need.

## Chapter 3

## Hypotheses

Hypothesis 1: Discrepancy in scores between mother-son dyads and mother-daughter dyads*:*

1. It is hypothesized that there will be a significantly smaller discrepancy in scores between child-reported and maternal-reported QOL in female-female dyads than female-male dyads.
2. It is hypothesized that mother-son dyads affected by LQTS will have a higher discrepancy in scores between child coping skills and parental perception of QOL than mother-daughter dyads as assessed through the PedsQL Generic 4.0 Core Scales and the PedsQL 3.0 Cardiac Module child and parent-proxy measures for each subscale and cumulatively.

Hypothesis 2:

1) QOL outcomes based on gender: It is hypothesized that female children with LQTS will have higher scores than males on child-reported QOL measures, indicating that they perceive themselves as having a better QOL than males. Child-reported scores on the PedsQL Generic 4.0 Core Total, and the PedsQL 3.0 Cardiac Module will be compared across gender for each subscale and combined.

# Chapter 4

## Methods

### Overview

The current study focuses on mothers and children who have been affected by LQTS and is part of a larger study designed to examine quality of life, coping, and psychological wellbeing in children with LQTS and their primary caregivers (Felgoise & Vetter, 2007). The current study analyzed a secondary data set, which was extracted from the original study. The participants selected from the original data set were chosen in an effort to examine the discrepancy between mother-son and mother-daughter dyads. The limited psychosocial research on gender differences in coping led to a focus on discrepancies between these dyads that can be investigated further in future research.

#### Design and design justification.

There has been relatively little research conducted on gender differences in coping and differences in parent/child coping in the LQTS population. In order to evaluate the differences between mother-daughter and mother-son dyads within this population, a between-subjects retrospective design was chosen. This type of design provided the opportunity to evaluate the mother-son dyads in comparison to mother-daughter dyads. The study assessed QOL of children by both child self-report and parent-proxy report and the QOL of children themselves. Such measurements provided a more comprehensive evaluation of quality of life of families as a whole. The analysis provided information about the interaction within mother-daughter and mother-son dyads and

about male and female children in the LQTS population, which has the potential to inform other groups of mothers and children managing chronic illness.

**Participants.**

For the present study, 35 mother-child dyads were assessed, 16 mother-son dyads and 19 mother-daughter dyads. Twenty-six female children and 19 male children were assessed.

**Inclusion and exclusion justification.**

English-speaking children ages 5-19 with LQTS attending one of two cardiology clinics affiliated with Children's Hospital of Philadelphia (CHOP) were recruited for participation with parental permission. Informed consent was obtained from their parents, and assent was obtained from the children. All children/adolescents/young adults between the ages of 5 and 19 in the same family were invited to participate in the study.

The sample for the current study evaluated the data of children from each family that consented. If they had more than one child with LQTS, parents were asked to complete the parent-proxy measures and personal information questionnaire for each child with LQTS. The present study examined sons and daughters and their mothers.

**Parent/Guardian Inclusion Criteria.**

One parent/guardian for each child with LQTS was recruited for participation if they met the following criteria: the parent was 18 years or older and was legally capable of giving informed consent; the parent could read/write English, as indicated by their ability to read and sign the informed consent form; the parent was willing to give

informed consent for him/herself and child with LQTS; the parent self-identified as the primary caregiver for their child with LQTS. “Primary caregivers” were defined as the parent who assumed primary responsibility for the child’s basic needs (activities of daily living), the administration of medications, if relevant, emotional support, attending their child’scardiac appointments, and supervising homework or school activity participation.

The secondary data set chosen for the present study considered only female primary caregivers.

**Parent Exclusion Criteria.**

Parents were excluded for the following reasons: if they were unable or unwilling to provide informed consent; were unable to read the questionnaires; or were unable to have the questionnaires read to them. For the secondary data sample set, fathers were excluded.

**Child Exclusion Criteria.**

Children with any major medical illness other than LQTS or cognitive impairment (i.e., developmental disabilities), as indicated by medical history, were excluded from this study due to the potential confounding that a dual diagnoses would impose. If the treating cardiologist identified the child as a member of a family that had recently (within 6 months) experienced an LQTS-related death, the child/parent was not contacted for participation. Children who came to clinic because of an acute change in their status or illness were excluded. Individuals who were not English speakers were excluded, because comparable instrumentation was not available for other languages.

**Recruitment**

A cardiologist at CHOP asked patients if they would be interested in participating in a study examining QOL and LQTS. Those who agreed were provided with an explanation of the Informed Consent/Assent.

Patients who qualified for the study were offered two participation options: (1) To employ a 90-minute time slot prior to their next regularly scheduled outpatient appointment to sign consent forms and assent forms and to complete questionnaires. Participants were able to finish questionnaires at home, if they were unable to complete the questionnaires in the clinic conveniently. A self-addressed envelope was provided to them for return of the questionnaires. Participants were asked to complete and return the questionnaires within one week. (2) Alternatively, participants could choose to provide consent and assent during their regularly scheduled outpatient appointment, and could complete the questionnaires at home and return them via a supplied stamped self-addressed envelope within one week. In either case, if materials were returned by mail, the questionnaires had only the participants' study number on them, and no names or other identifying information were to be enclosed in the packet. Participants received a movie ticket for each participant after completed questionnaires were received.

**Suicide Screening.**

Although it is unlikely that participants would experience emotional discomfort from discussing topics related to the QOL of children or adolescents who have heart disease, suicide screening was implemented. Items related to harm and/or suicidality on study questionnaires were targeted for review by the research assistants. Items targeted were the following: Beck Depression Inventory-II --Question # 2, Pessimism; Question #

9, Suicidal thoughts or wishes. Beck Youth Inventory--Question #44:, I wish I were dead. Child Depression Inventory--Question # 9, Suicidal thoughts or wishes.

In addition to the suicide screening, a referral list was provided to all patients that included psychology referrals, online support groups, and informational websites; each participant was also offered more detailed information sheets on anxiety and depression at the time of consent. Every study participant received a resource list of qualified mental health professionals, and was offered additional pamphlets explaining anxiety and depression, conditions commonly encountered in this population. The list provided a starting point for those individuals with self-disclosed or observable stress, in addition to those who did not want to admit they were suffering from mental health issues.

**Measures**

The PedsQL Measurement Model was created to measure health-related quality of life (HRQOL) in children and adolescents, and the PedsQL 4.0 Generic Core Scales was designed to integrate with the PedsQl Disease-Specific Modules (Varni, Sherman, Burwinkle, Dickinson, & Dixon, 2004). The Pediatric Quality of Life Inventory Version 4.0 measures QOL according to the World Health Organization's definition, with an additional school functioning scale (Varni, Sherman, Burwinkle, Dickinson, & Dixon, 2004). It was developed as a modular approach to measuring pediatric HRQOL for children ages 2-18, with generic core scales and disease-specific modules (Varni et al., 1999). The PedsQl 4.0 Generic Core Scales and the PedsQl 3.0 Cardiac Module child self-report and parent-proxy measures were used in the present study. The PedsQL is a brief, standardized generic assessment instrument that systematically assesses patients'

and parents' perceptions of health-related quality of life (HRQOL) in pediatric patients with chronic health conditions, using pediatric cancer as the model. HRQOL is defined in terms of the impact of disease and treatment on an individual's physical, psychological, and social functioning, and by disease/treatment-specific symptoms. In regard to HRQOL measurement, it has been found that individuals in the best possible physical and emotional state among those with their medical condition have the best possibility of achieving a high quality of life.

The 23-item PedsQL 4.0 generic core scales encompass physical functioning (8 items), emotional functioning (5 items), social functioning (5 items), and school functioning (5 items). The Generic Core Scales are comprised of parallel child self-report and parent proxy-report formats. Child self-report includes ages 5-7, 8-12 and 13-18. The parent proxy-report forms are parallel to the child self-report forms, and designed to assess the parent's perceptions of their child's HRQOL. The items in each are fundamentally identical, diverging in developmentally appropriate language and use of the first or third person. The instructions ask the individual how much of a problem each item has been during the past month. A 5-point Likert scale is implemented across child self-report for ages 8-18 and parent proxy-report (0 = *never a problem*; 1 = almost never *a problem;* 2 = *sometimes a problem;* 3 = *often a problem*; 4 = *almost always a problem*). For the young child self-report (ages 5-7), the Likert scale is rephrased and simplified to a 3-point scale (0 = *not at all a problem*; 2 = *sometimes a problem;* 4 = *often a problem*), with each response connected to a happy-sad faces scale (Varni et al., 2001). Items are reverse-scored and linearly transformed to a 0-100 scale (0 = 100, 1 = 75, 2 = 50, 4 = 0), so that higher PedsQL 4.0 scores indicate higher HRQOL.

The Physical Health Summary Score (8 items) consists of the Physical Functioning Scale. The Psychosocial Health Summary Score (15 items) is computed by adding the other items answered in the Emotional, Social, and School Functioning Scales and dividing it by the number answered. In a study by Varni et al. (2001), the PedsQL was administered to 209 children and 269 parents. The following data emerged for healthy individuals: a mean of 83.00 for total score, 84.41 for physical health, and 82.38 for psychosocial health (Varni et al., 2002). For individuals with cardiac conditions in Class II (those with symptoms present with greater than usual exertion or prescribed limitations), the following data emerged: a mean of 75.88 for total score, 78.73 for physical health, and 74.31 for psychosocial health (Varni et al., 2002). Parent-proxy measures utilized with parents of healthy children were found to have a mean of 87.61 for total score, 89.32 for physical health, and 86.58 for psychosocial health, but parent-proxy measures used with parents of children with cardiac conditions in Class II showed a mean of 80.08 for total score, 82.77 for physical health, and 78.46 for psychosocial health.

The cardiac disease-specific module's development was based on methodology previously utilized by Varni and colleagues to develop disease-specific modules for asthma, arthritis, diabetes, and cancer. The measure was based upon studies of the cardiac-specific health-related quality of life in empirical literature and discussions with pediatric cardiology healthcare providers. Wording of items from other disease-specific modules was used in the measure as well (Uzark, Jones, Burwinkle, & Varni, 2003). The child and parent reports of the PedsQL 3.0 Cardiac Module are composed of 27 items comprising 6 dimensions. It has 5 scales related to symptoms (7 items), perceived physical appearance (3 items), treatment anxiety (4 items), cognitive problems (5 items),

and communication (3 items) for parent proxy-reports and children 8–18 years. The communication scale was not included for young children who did not have the cognitive or language resources available needed to verbalize questions and concepts about the heart. An additional treatment barriers scale, the 6th scale, (5 items) is a part of the module used to measure compliance issues in subjects taking cardiac medications (Uzark, Jones, Burwinkle, & Varni, 2003). Items are reverse-scored and linearly transformed to a 0-100 scale (0 = 100, 1 = 75, 2 = 50, 4 = 0), so that higher PedsQL 3.0 Cardiac Module scores indicate higher HRQOL.

The reliability and validity of the PedsQL generic has been shown in healthy and patient populations with >30000 children, adolescents and their parents around the world. Both reliability and validity have been confirmed (Varni, Seid, & Rode, 1999). The internal consistency for all items has yielded patient' report ([alpha] = .93) and parents' reports ([alpha] = .93) (Varni et al., 1999). Cronbach's alphas suggested that the PedsQL is internally consistent and measures a unitary construct (Varni et al., 1999). The Psychological and Social Functioning Scales have been found to correlate with standardized scales of emotional distress, compliance perception, and social functioning (Varni et al., 1999).

**Procedure**

Research material obtained from individuals consisted of completed inventories and questionnaires. All written consent/assent forms were separated from study materials upon receipt and kept in a separate file from patient study materials. All materials (consents, assents, and completed questionnaires) were locked in a file cabinet at PCOM.

The secondary data set that was analyzed only utilized the PedsQL child and parent-proxy measures from each completed inventory.

At PCOM, all study information was housed in individual hanging file folders in a locked cabinet, and in a secure and de-identified, password-protected study database. Informed consent forms were kept in a separate file from data materials to ensure that data was de-identified during filing, scoring, data entry, and analysis. The study files and the study database did not contain subjects' names; instead, a unique study number was assigned to each patient/parent pair and used to identify information specific to each de-identified participant.

**Security.**

The study database had no identifiers and was backed up on two jump drives and on PCOM's secure network server.

**Statistical Analysis**

To test hypothesis 1, a multivariate analysis of variance (MANOVA) was conducted to assess if there were differences between mother-son dyads and mother-daughter dyads on the PedsQl Generic 4.0 Core Scale and the PedsQL 3.0 Cardiac Module child and parent-proxy measures. A one-factor MANOVA was used to perform comparisons when multiple dependent variables needed to be analyzed together. In this case, gender was the independent variable, and subscales across the PedsQl Generic 4.0 Core Total child and parent-proxy measures were the dependent variables. A MANOVA was used to see the dependent variable differences between groups. Discrepancy, also

called cross-informant variance, between mother-son and mother-daughter perception and self-reported QOL was compared across dyads and subscales. Differences in QOL were compared across the children's gender.. The one-factor MANOVA helped to identify differences between mother-son and mother-daughter dyads.

To test hypothesis 2, a *t*-test was conducted to compare Quality of Life between males and females on the PedsQL 4.0 Generic Core Scales and the PedsQl 3.0 Cardiac Module child measures, across subscales and combined.

# Chapter 5

## Results

### Study Population

Data were collected from 19 boys with LQTS, 26 girls with LQTS, and their mothers. Some participants did not complete the measures fully or did not return the measures, which was reported in the subsequent data analyses.

Of the 45 mothers in the study, 33 filled out measures indicating that they were themselves diagnosed with LQTS, and 16 of those 33 identified as being symptomatic. The range for mothers' ages fell between 32 and 58 years old, with a mean of 45. In this study, 26 mothers reported having religious beliefs.

The personal information questionnaire (PIQ) asked the age when the child was first diagnosed with LQTS, and when they first began treatment for LQTS; 5 children were diagnosed and treated from birth to age 3, 4 children were diagnosed and treated at ages 5 and 6, 6 were diagnosed and treated from ages 8 to 12, and 4 were diagnosed and treated at ages 13 to 17. The data from the PIQ revealed that 9 children were previously diagnosed with other mental health or medical problems: 4 children with asthma, one child with depression, one with ADD, one with Dyslexia, one with OCD tendencies, and one with bipolar disorder and Type1 diabetes. In regard to their first symptomatic episode, 3 children fainted, one child kept falling asleep in school, and 4 did not specify. For frequency of LQTS symptoms, 17 children identified as asymptomatic, 2 children identified as having symptoms 1-3 times, and one had symptoms monthly. The PIQ data indicated that 3 children had pacemakers at the time of the study, one had both, and the

rest had neither. For frequency of cardiology appointments, 2 children went every 3 months, 6 went twice a year, and 12 went yearly.

The following table shows the sample size, mean, and standard deviation of male children with LQTS who completed the PedsQl 4.0 Generic Core Scale Child measure. Average scores ranged from 71.00 to 93.48 as opposed to average scores of healthy children, 83 to 100. Boys ages 8-12 were found to have the lowest scale score of 71.00 in the Emotional Functioning subscale. Boys ages 8-12 also scored lower than the adolescents or young children in the school functioning and psychosocial health summary subscales, 72.5 and 75.00, respectively. Adolescent boys 13-19 were found to have average scores of 75.00 on the Emotional Functioning subscale and 73.75 on the Psychosocial Health Summary Score. Both children and adolescents had total scale scores under 80. Overall, the lowest scores for all groups were found on the Emotional Functioning subscale, the Social Functioning subscale, and the Psychosocial Healthy Summary subscale. The data on young children were underpowered with a sample size of one; therefore, the data did not contribute to the overall analysis.

*Table 1. PedsQL 4.0 Generic Core Scales*

| Children: Male | 5-7 | | | 8-12 | | | 13-19 | | |
|---|---|---|---|---|---|---|---|---|---|
| Dx w/ LQTS | N | X | SD | N | X | SD | N | X | SD |
| Physical Functioning | 1 | 81.25 | | 10 | 85.32 | 7.66 | 8 | 82.81 | 16.37 |
| Emotional Functioning | 1 | 100 | | 10 | 71.00 | 16.12 | 8 | 75.00 | 20.35 |
| Social Functioning | 1 | 100 | | 10 | 81.5 | 15.28 | 8 | 84.38 | 15.68 |
| School Functioning | 1 | 100 | | 10 | 72.5 | 12.53 | 8 | 73.75 | 21.99 |
| Psychosocial Health Summary | 1 | 100 | | 10 | 75.00 | 14.08 | 8 | 77.50 | 14.53 |
| Physical Health Summary | 1 | 81.25 | | 10 | 86.32 | 8.16 | 8 | 82.81 | 16.37 |
| Total Scale | 1 | 93.48 | | 10 | 78.59 | 10.77 | 8 | 79.35 | 12.72 |

The following table shows the sample size, mean, and standard deviation of female children with LQTS who completed the PedsQl 4.0 Generic Core Scale Child measure. Average scores ranged from 60.71 for girls 8-12 on the Emotional Functioning subscale to 85.42 for young girls on the Physical Functioning subscale and Physical Health summary subscale. Higher average scores connected to physical aspects of LQTS may be explained by the typical asymptomatic nature of LQTS. The lowest average scores for all groups were found on the Emotional Functioning Subscales. Total scale scores ranged from 68.63 to 76.81, with girls in the 8-12 age group having lower average

scores in all categories especially as compared to healthy children, who score in the 83 to 100 range.

*Table 2. PedsQL 4.0 Generic Core Scales*

| Children: Female | 5-7 | | | 8-12 | | | 13-19 | | |
|---|---|---|---|---|---|---|---|---|---|
| Dx w/ LQTS | N | X | SD | N | X | SD | N | X | SD |
| Physical Functioning | 3 | 85.42 | 13.01 | 7 | 73.66 | 21.86 | 16 | 83.32 | 13.89 |
| Emotional Functioning | 3 | 66.67 | 25.17 | 7 | 60.71 | 23.70 | 16 | 66.56 | 25.01 |
| Social Functioning | 3 | 73.33 | 15.28 | 7 | 68.57 | 28.83 | 16 | 82.5 | 18.71 |
| School Functioning | 3 | 76.67 | 15.28 | 7 | 68.57 | 25.93 | 16 | 76.56 | 21.27 |
| Psychosocial Health Summary | 3 | 72.22 | 15.75 | 7 | 65.95 | 25.26 | 16 | 74.48 | 18.82 |
| Physical Health Summary | 3 | 85.42 | 13.01 | 7 | 73.66 | 21.86 | 16 | 83.32 | 13.89 |
| Total Scale | 3 | 76.81 | 14.47 | 7 | 68.63 | 23.83 | 16 | 77.46 | 17.20 |

It was found that male children scored higher than females overall on the PedsQL 4.0 Generic core scales measure. For male and female children, 5-7, the mean total scores were 93.48 and 76.81, respectively. The only subscales where females had higher scores were physical functioning, 85.42 compared to 81.25, and physical health summary, 84.42 compared to 81.25. For male and female children, 8-12, the mean total scores were 78.59 and 68.63, respectively. Male children 8-12 scored higher on every subscale. For male and female adolescents, 13-19, the mean total scores were 79.35 and 77.46, respectively.

The only subscales in which males had higher scores were physical functioning, 83.32 compared to 82.81 school functioning, 76.56 compared to 73.75, and physical health summary, 83.32 compared to 82.81. The mean of total scores for all age groups was 83.81 for male children, and 74.3 for female children, markedly lower than the average healthy child, 83 to 100. These data were contrary to the hypothesis, but may be indicative of females being more forthcoming or honest about their responses than males.

The following table shows the sample size, mean, and standard deviation of mothers of male children with LQTS who completed the PedsQl 4.0 Generic Core Scale Parent Proxy measure. Average scores ranged from 70.5 to 100, with the lowest average attributed to parents of male children 8-12 on the Social Functioning subscale. The parent-proxy measure was underpowered for young males 5-7, with only one parent measure analyzed. With parents of both children and adolescents, higher scale scores were found on the Physical Functioning and Physical Health Summary scale score, with lower average scores found on the Emotional, Social, School, and Psychosocial health summary subscales. Parents of male children 8-12 were found to have the lowest total scale score, 73.59.

*Table 3. PedsQL 4.0 Generic Core Scales*

| Parent: Male | 5-7 | | 8-12 | | | 13-19 | | | Total | |
|---|---|---|---|---|---|---|---|---|---|---|
| Child Dx w/ LQTS | N | X | N | X | SD | N | X | SD | X | SD |
| Physical Functioning | 1 | 100 | 10 | 76.94 | 25.16 | 7 | 82.14 | 19.15 | | |
| Emotional Functioning | 1 | 90 | 10 | 72.5 | 17.04 | 7 | 71.43 | 18.19 | | |
| Social Functioning | 1 | 100 | 10 | 70.5 | 20.88 | 7 | 77.14 | 23.6 | | |
| School Functioning | 1 | 100 | 10 | 72.17 | 16.63 | 7 | 74.29 | 26.52 | | |
| Psychosocial Health Summary | 1 | 96.67 | 10 | 73.33 | 15.28 | 7 | 74.29 | 20.04 | | |
| Physical Health Summary | 1 | 100 | 10 | 74.64 | 25.39 | 7 | 82.14 | 19.15 | | |
| Total Scale Score | 1 | 97.83 | 10 | 73.59 | 16.72 | 7 | 77.02 | 19.18 | | |

The following table shows the sample size, mean, and standard deviation of mothers of female children with LQTS who completed the PedsQl 4.0 Generic Core Scale Parent Proxy measure. Average scores ranged from 56.25 to 90, with the lowest averages attributed to parents of female children 8-12 on the Physical Functioning, Physical Health Summary, and Emotional Functioning subscales. The parent-proxy measure was underpowered for the young females 5-7, with only two parent measures analyzed. However, there were only 5 parents of female children ages 8-12. With parents

of adolescents, higher scale scores were found on the Physical Functioning and Physical Health Summary subscale scores, and lower average scores found on the Emotional, Social, School, and Psychosocial healthy summary subscales. Parents of female children 8-12 were found to have the lowest total scale score of 60.87, compared to adolescents and small children, 81.71 and 84.24, respectively. The standard deviations for the subscales for the parents of the 8-12 age group were significantly larger than the standard deviations for the other age groups on the generic core scale measures.

*Table 4. PedsQL 4.0 Generic Core Scales*

| Parent: Female | 5-7 | | | 8-12 | | | 13-19 | | |
|---|---|---|---|---|---|---|---|---|---|
| Child Dx w/ LQTS | N | X | SD | N | X | SD | N | X | SD |
| Physical Functioning | 2 | 84.38 | 13.26 | 5 | 56.25 | 34.45 | 15 | 88.75 | 10.42 |
| Emotional Functioning | 2 | 75.00 | 7.07 | 5 | 57 | 33.65 | 15 | 72.33 | 21.20 |
| Social Functioning | 2 | 90.00 | 14.14 | 5 | 63 | 20.19 | 15 | 83.33 | 17.39 |
| School Functioning | 2 | 87.50 | 17.68 | 5 | 70 | 42.28 | 15 | 71.00 | 26.06 |
| Psychosocial Health Summary | 2 | 89.17 | 3.53 | 5 | 63.33 | 30.11 | 15 | 75.55 | 19.1 |
| Physical Health Summary | 2 | 84.38 | 13.26 | 5 | 56.25 | 34.45 | 15 | 88.75 | 10.42 |
| Total Scale Score | 2 | 84.24 | 2.31 | 5 | 60.87 | 28.85 | 15 | 81.71 | 12.28 |

It was found that mothers of male children 8-12, and young children, 5-7, identified higher QOL than mothers of females on the PedsQL 4.0 Generic core scales measure. Mothers of adolescents 13-19 did, however, report higher QOL for female adolescents than male adolescents. For mothers of male and female children 5-7, the mean total scale scores were 97.83 and 84.24 , respectively. For mothers of male and female children 8-12, the mean total scores were 73.59 and 60.87, respectively. Parents of male children 5-7 and 8-12 gave higher QOL perception scores on every subscale. For mothers of male and female adolescents 13-19, the mean total scores were 77.02 and 81.71, respectively. The only measure for which mothers of males gave their sons higher scores was school functioning, 74.29 compared to 71.00. The mean of total scores for all age groups was 82.81 for mothers of male children and 75.61 for mothers of female children.

The following table shows the sample size, mean, and standard deviation of male children with LQTS who completed the PedsQl 3.0 Cardiac Module child measure. Average scores ranged from 69.5 to 95.31, with the lowest averages attributed to male children 8-12 on the Cognitive Problems and Communication subscales. For both male children and adolescents, higher scores were found on the Treatment Anxiety and Perceived Physical Appearance subscales, with lower average scores found on the Heart Problems and Treatment and Cognitive Problems subscales. The largest discrepancies in scores were found on the Cognitive Problems and Communication subscales, with male children 8-12 scoring lower than male adolescents 13-19.

*Table 5. PedsQL 3.0 Cardiac Module*

| Children: Male | 5-7 | | | 8-12 | | | 13-19 | | |
|---|---|---|---|---|---|---|---|---|---|
| Dx w/ LQTS | N | X | SD | N | X | SD | N | X | SD |
| Heart Problems and Tx | 0 | | | 10 | 76.07 | 10.38 | 8 | 80.36 | 8.32 |
| Treatment II | 0 | | | 10 | 90.00 | 13.12 | 8 | 84.38 | 20.08 |
| Perceived Physical Appearance | 0 | | | 10 | 88.75 | 9.63 | 8 | 88.75 | 17.83 |
| Treatment Anxiety | 0 | | | 10 | 90.00 | 11.49 | 8 | 95.31 | 13.26 |
| Cognitive Problems | 0 | | | 10 | 69.50 | 17.23 | 7 | 83.57 | 9.88 |
| Communication | 0 | | | 10 | 72.5 | 29.41 | 7 | 84.52 | 20.65 |
| Total | | | | 10 | 81.14 | | 7 | 86.15 | |

It was found that male children scored higher than females children overall on the PedsQL 3.0 Cardiac Module measure. The only measures in which females had higher scores were treatment barriers, 92.50 compared to 90, and cognitive problems, 75.83 compared to 69.50. For male and female children, 8-12, the mean of the subscales was 81.14 and 75.83, respectively. For males and female adolescents, 13-19, the mean of the subscales were 86.15 and 76.43, respectively. This data was contrary to the hypothesis, but may indicate that females are more forthcoming or honest about their QOL in contrast to males. Young children were unable to be compared due to lack of data for boys 5-7.

The following table shows the sample size, mean, and standard deviation of female children with LQTS who completed the PedsQl 3.0 Cardiac Module child measure. Average scores ranged from 66.67 to 100, with the lowest averages attributed to

female children 5-7 and 8-12 on the Communication subscale, and adolescent females 13-19 on the Treatment Anxiety subscale. Higher scale scores were found on the Perceived Physical Appearance and Cognitive Problems subscales for young female children 5-7 and the Treatment II subscale for female children. The majority of the scores listed on this table fell below 80, indicating a lower level of self-perceived cardiac-related QOL.

*Table 6. PedsQL 3.0 Cardiac Module*

| Children: Female | 5-7 | | | 8-12 | | | 13-19 | | |
|---|---|---|---|---|---|---|---|---|---|
| Dx w/ LQTS | N | X | SD | N | X | SD | N | X | SD |
| Heart Problems and Tx | 3 | 88.1 | 10.91 | 6 | 74.4 | 15.88 | 17 | 75.21 | 15.02 |
| Treatment II | 3 | 83.33 | 28.87 | 6 | 92.5 | 6.12 | 17 | 83.82 | 20.35 |
| Perceived Physical Appearance | 3 | 100 | .00 | 6 | 83.33 | 14.91 | 17 | 78.43 | 30.49 |
| Treatment Anxiety | 3 | 79.17 | 19.1 | 6 | 73.96 | 39.21 | 17 | 68.75 | 34.30 |
| Cognitive Problems | 3 | 90 | 10.00 | 6 | 75.83 | 21.08 | 17 | 77.35 | 17.86 |
| Communication | 3 | 66.67 | 16.67 | 6 | 68.1 | 30.92 | 17 | 75.00 | 30.76 |
| Total | 3 | 84.55 | | 6 | 78.02 | | 17 | 76.43 | |

The following table shows the sample size, mean, and standard deviation of mothers of males with LQTS who completed the PedsQl 3.0 Cardiac Module parent proxy measure. Average scores ranged from 60 to 100, with the lowest averages

attributed to parents of male children 8-12, and male adolescents 13-19, on the Communication and Cognitive Problems subscales. The data on parents of young children were underpowered with a sample size of one, and therefore the data did not add much to the overall analysis. Higher scale scores were found on the Treatment II and Heart Problems and Treatment subscales for parents of male children. The data revealed that parents of children and adolescent males identified deficiencies in their children QOL connected to Cognitive Problems and Communication.

*Table 7. PedsQL 3.0 Cardiac Module*

| Parent: Male | 5-7 | | | 8-12 | | | 13-19 | | |
|---|---|---|---|---|---|---|---|---|---|
| Child Dx w/ LQTS | N | X | SD | N | X | SD | N | X | SD |
| Heart Problems and Tx | 1 | 96.43 | | 10 | 87.50 | 12.28 | 7 | 87.25 | 15.13 |
| Treatment II | 1 | 100 | | 9 | 93.70 | 7.67 | 7 | 84.29 | 12.05 |
| Perceived Physical Appearance | 0 | | | 10 | 81.04 | 20.66 | 7 | 91.67 | 18.63 |
| Treatment Anxiety | 1 | 81.25 | | 10 | 71.75 | 22.59 | 7 | 77.68 | 28.84 |
| Cognitive Problems | 1 | 100 | | 10 | 62.00 | 31.90 | 7 | 60 | 24.66 |
| Communication | 1 | 100 | | 10 | 65.00 | 17.92 | 7 | 65.48 | 32.43 |
| Total | 1 | 95.54 | | 10 | 76.83 | | 7 | 77.73 | |

The following table shows the sample size, mean, and standard deviation for mothers of females with LQTS who completed the PedsQl 3.0 Cardiac Module parent proxy measure. Average scores ranged from 60 to 95.87, with the lowest averages

attributed to parents of female children 8-12 on the Heart Problems and Treatment, Treatment Anxiety, and Cognitive Problems subscales. Higher scale scores were found on Treatment II, Heart Problems and treatment, and Perceived Physical Appearance subscales for parents of female young children. This data may not add much to the analysis, since there was an N of only 2. The data revealed that parents of male children reported problems for their children in almost all of the Cardiac QOL domains.

*Table 8. PedsQL 3.0 Cardiac Module*

| Parent: Female | 5-7 | | | 8-12 | | | 13-19 | | |
|---|---|---|---|---|---|---|---|---|---|
| Child Dx w/ LQTS | N | X | SD | N | X | SD | N | X | SD |
| Heart Problems and Tx | 2 | 91.08 | 2.52 | 5 | 60 | 23.20 | 15 | 86.90 | 12.85 |
| Treatment II | 2 | 91.67 | 11.79 | 5 | 87 | 18.25 | 15 | 89.1 | 8.82 |
| Perceived Physical Appearance | 2 | 95.84 | 5.89 | 5 | 76.67 | 22.36 | 15 | 77.78 | 29.32 |
| Treatment Anxiety | 2 | 75.00 | .00 | 5 | 62.5 | 15.31 | 15 | 73.42 | 29.86 |
| Cognitive Problems | 2 | 77.50 | 3.54 | 5 | 61 | 39.27 | 15 | 68.67 | 20.91 |
| Communication | 2 | 87.50 | 17.68 | 5 | 70 | 29.23 | 15 | 81.11 | 23.88 |
| Total | 2 | 86.43 | | 5 | 69.53 | | 15 | 79.5 | |

It was found that mothers of male children 5-7 and 8-12 reported higher QOL than mothers of females on the majority of the scales on PedsQl 3.0 Cardiac Module measure. Mothers of adolescents 13-19, however, reported higher QOL overall for female adolescents than male adolescents. For mothers of young male and female children 5-7, the mean total scores were 95.54 and 86.43, respectively. For mothers of males and female children, 8-12, the mean total scores were 76.83 and 69.5, respectively. Parents of male children 5-7 gave higher QOL perception scores than mothers of female children on every subscale. The only scale in which parental perception of quality of life was higher for mothers of female children was on the communication subscale for girls 8-12, 70.00 for mothers of girls and 65.00 for mothers of boys. For mothers of male and female adolescents, 13-19, the mean total scores were 77.3 and 79, respectively. The mean total scores for all age groups was 83.37 for mothers of male children and 78.49 for mothers of female children.

The following table shows the sample size, mean, and standard deviation for mothers of male and female children of all ages with LQTS who completed the PedsQL 4.0 Generic Core Scales parent proxy measure and the PedsQL 3.0 Cardiac Module parent proxy measure. Average scores ranged from 65.14 to 89.69, with the lowest averages found in a parent's perception of her child's emotional functioning, 71.71, and in a parent's perceptions of her child's cognitive problems, 65.14, and her child's treatment anxiety, 72.50. Higher scale scores were attributed to parents' perceptions of their child's Heart Problems and Treatment, 85.00, Treatment barriers, 89.69, and Physical functioning, 81.76. The data revealed that parents reported difficulties in their

children in more than half of all QOL domains, Generic and Cardiac, as evidenced by average scores lower than 80 on more than half of the subscales.

*Table 9. PedsQL 4.0 Generic Core Scales and PedsQL 3.0 Cardiac Module*

| | All Ages | | |
|---|---|---|---|
| Parents of All Children Dx w/ LQTS | N | X | SD |
| Physical Functioning | 35 | 81.72 | 21.40 |
| Emotional Functioning | 35 | 71.71 | 18.15 |
| Social Functioning | 35 | 77.43 | 19.72 |
| School Functioning | 35 | 74.62 | 22.55 |
| Psychosocial Health Summary | 35 | 75.05 | 17.02 |
| Physical Health Summary | 35 | 81.06 | 21.65 |
| Total Scale Score | 35 | 77.75 | 15.65 |
| Heart Problems and Tx | 35 | 85.00 | 14.20 |
| Treatment II | 34 | 89.70 | 9.28 |
| Perceived Physical Appearance | 35 | 83.16 | 23.22 |
| Treatment Anxiety | 35 | 72.5 | 25.27 |
| Cognitive Problems | 35 | 65.14 | 24.57 |
| Communication | 35 | 74.29 | 24.12 |

The following table shows the sample size, mean, and standard deviation of male and female children all ages diagnosed with LQTS, who completed the PedsQL 4.0 Generic Core Scales measure and the PedsQL 3.0 Cardiac Module measure. Average

scores ranged from 69.76 to 86.5, with the lowest averages found in a child's perception of their emotional functioning, 69.76, and a child's perception of their communication, 73.93. Higher scale scores were attributed to a child's perception of their Treatment barriers, 86.50, Physical functioning, 82.97, and Perceived Physical Appearance, 83.27. The data revealed that children reported difficulties in more than half of all QOL domains, Generic and Cardiac, as evidenced by average scores lower than 80 on more than half of the subscales.

*Table 10. PedsQL 4.0 Generic Core Scales and PedsQL 3.0 Cardiac Module*

| | All Ages | | |
|---|---|---|---|
| All Children Dx w/ LQTS | N | X | SD |
| Physical Functioning | 41 | 82.97 | 14.29 |
| Emotional Functioning | 41 | 69.76 | 21.09 |
| Social Functioning | 41 | 81.46 | 18.48 |
| School Functioning | 41 | 75.12 | 18.86 |
| Psychosocial Health Summary | 41 | 75.12 | 16.97 |
| Physical Health Summary | 41 | 83.22 | 14.40 |
| Total Scale Score | 41 | 77.82 | 15.29 |
| Heart Problems and Tx | 40 | 76.43 | 13.26 |
| Treatment II | 40 | 86.50 | 17.25 |
| Perceived Physical Appearance | 40 | 83.27 | 22.48 |
| Treatment Anxiety | 40 | 79.69 | 29.58 |
| Cognitive Problems | 39 | 76.03 | 17.29 |
| Communication | 39 | 73.93 | 28.11 |

The following table shows the sample size, mean, and standard deviation of the differences between mothers' perceptions of the QOL of their female children, and their child's perception of their own QOL as identified on the PedsQL 4.0 Generic Core Scales child and parent-proxy measure and the PedsQL 3.0 Cardiac Module child and parent-proxy measure. Average difference scores ranged from 11.84 to -5.26, with the greatest

differences in parent and child perceptions of their Communication, 11.84, and in parent and child perceptions of Cognitive Problems -5.52. Child scores were subtracted from Parent scores. Therefore, negative scores indicate where parents perceived their female child's QOL as worse than the child's perceptions across QOL scales, and positive scores indicate where parents perceived their female child's QOL as better than the child's perceptions. The data revealed that parents perceived their female children as having higher QOL in all QOL domains, Generic and Cardiac, except for Cognitive problems, as evidenced by positive average difference scores on all of the subscales except Cognitive problems. Difference scores did not reach statistical significance level of .05 for any of the subscales.

*Table 10. PedsQL 4.0 Generic Core Scales and PedsQL 3.0 Cardiac Module*

All Ages

Differences Between Mothers and Females

| All Ages Child Dx w/ LQTS | N | X | SD |
|---|---|---|---|
| Physical Functioning | 19 | 1.05 | 13.71 |
| Emotional Functioning | 19 | 5.52 | 16.99 |
| Social Functioning | 19 | 1.84 | 21.87 |
| School Functioning | 19 | .26 | 20.38 |
| Psychosocial Health Summary | 19 | 3.15 | 16.15 |
| Physical Health Summary | 19 | 1.05 | 13.71 |
| Total Scale Score | 19 | 3.74 | 16.01 |
| Heart Problems and Tx | 19 | 7.14 | 16.28 |
| Treatment II | 19 | 5.59 | 19.67 |
| Perceived Physical Appearance | 19 | 2.63 | 22.74 |
| Treatment Anxiety | 19 | 5.33 | 25.91 |
| Cognitive Problems | 19 | -5.26 | 18.59 |
| Communication | 19 | 11.84 | 34.17 |

The following table shows the sample size, mean, and standard deviation of the differences between mothers' perceptions of the QOL of their male children, and their child's perception of their own QOL as reported on the PedsQL 4.0 Generic Core Scales child and parent-proxy measure and the PedsQL 3.0 Cardiac Module child and parent-

proxy measure. Average difference scores ranged from -19.6 to 9.15, with the greatest differences in parent and child perceptions of their Treatment Anxiety, -19.6, and parent and child perceptions of their Heart Problems, 9.15. Child scores were subtracted from Parent scores. Therefore, negative scores indicate where parents perceived their male child's QOL as worse than the child perceptions across QOL scales, and positive scores indicate where parents perceived their male child's QOL as better than the child's perceptions across QOL subscales. The data revealed that parents perceived their male children as having lower QOL on all QOL domains, Generic and Cardiac, except School Functioning and Heart Problems, as evidenced by negative average difference scores on all of the subscales besides School Functioning and Heart Problems. Difference scores did not reach statistical significance level of .05 on any of the subscales.

*Table 11. PedsQL 4.0 Generic Core Scales and PedsQL 3.0 Cardiac Module*

| | All Ages | | |
|---|---|---|---|
| Differences Between Mothers and Males | | | |
| All Ages Child Dx w/ LQTS | N | X | SD |
| Physical Functioning | 16 | -6.02 | 24.60 |
| Emotional Functioning | 16 | -.63 | 19.91 |
| Social Functioning | 16 | -9.38 | 17.78 |
| School Functioning | 16 | 2.29 | 24.33 |
| Psychosocial Health Summary | 16 | -1.46 | 16.43 |
| Physical Health Summary | 16 | -8.05 | 24.79 |
| Total Scale Score | 16 | -3.67 | 15.18 |
| Heart Problems and Tx | 16 | 9.15 | 14.34 |
| Treatment II | 15 | -.44 | 9.97 |
| Perceived Physical Appearance | 16 | -1.28 | 16.26 |
| Treatment Anxiety | 16 | -19.6 | 22.72 |
| Cognitive Problems | 15 | -13.3 | 27.30 |
| Communication | 15 | -7.78 | 29.29 |

The following table shows the sample size, mean, and standard deviation of the differences in parents' perception of the QOL of their children, and their child's perception of their own QOL as reported on the PedsQL 4.0 Generic Core Scales child and parent-proxy measure and the PedsQL 3.0 Cardiac Module child and parent-proxy

measure. Average difference scores ranged from 8.06 to -8.82, with the greatest differences in parents' perceptions of their child's Heart Problems and Treatment, 8.06, and parents' perceptions of their child's Cognitive Problems, -8.82. Child scores were subtracted from Parent scores. Therefore, negative scores indicate where parents perceived their children's QOL as worse than the children's perceptions across QOL scales, and positive scores indicate where parents perceived their children's QOL as better than the children's perceptions. The data revealed that parents perceived their children as having higher QOL on more than half of all QOL domains, Generic and Cardiac, as evidenced by positive average difference scores on more than half of the subscales. Difference scores reached the level of statistical significance at .02 only on the treatment anxiety subscale. This indicated that boys had significant cross-informant variance for the Treatment Anxiety subscale (-19.6), as compared to girls (5.33).

*Table 12. PedsQL 4.0 Generic Core Scales and PedsQL 3.0 Cardiac Module*

| | All Ages | | |
|---|---|---|---|
| Differences Between All Ages, Both Genders, Child Dx w/ LQTS | N | X | SD |
| Physical Functioning | 35 | -2.18 | 19.47 |
| Emotional Functioning | 35 | 2.71 | 18.36 |
| Social Functioning | 35 | -3.29 | 20.61 |
| School Functioning | 35 | 1.19 | 21.95 |
| Psychosocial Health Summary | 35 | 1.05 | 16.21 |
| Physical Health Summary | 35 | -3.12 | 19.79 |
| Total Scale Score | 35 | .35 | 15.86 |
| Heart Problems and Tx | 35 | 8.06 | 15.23 |
| Treatment II | 34 | 2.93 | 16.21 |
| Perceived Physical Appearance | 35 | .85 | 19.86 |
| Treatment Anxiety | 35 | -6.07 | 27.24 |
| Cognitive Problems | 35 | -8.82 | 22.83 |
| Communication | 35 | 3.19 | 33.14 |

# Chapter 6

## Discussion

### Summary of findings

The author expected to find significant differences in QOL between males and females with LQ, as well as differences between QOL perceptions in mother-son and mother-daughter dyads. It was predicted that there would be significant differences between mother-son and mother-daughter dyads across each of the measures of interest: PedsQl Generic 4.0 Core total child and parent-proxy measures, and the PedsQl 3.0 Cardiac Module child and parent-proxy measures. The study was exploratory, and looking for preliminary differences between QOL in dyads. As previously discussed, there are few studies that examine these differences; therefore, claiming that differences are present at all can further the science, as well as validate previous studies that have claimed gender differences in similar populations.

Across both the Generic and Cardiac PedsQL measures, scores were consistently lower than scores of healthy children, which typically range from 83 to 100 (Varni et al., 2001). The majority of scores were found to be lower than 80, indicating lower QOL, and perceived QOL, for boys, girls, and parents. Considering that the majority of the study sample is asymptomatic, this finding indicates that merely having a diagnosis of LQTS could be reason enough for marked psychosocial stress.

Hypothesis 1 was supported in part, as the Treatment Anxiety subscale analysis scores indicated that the Treatment Anxiety subscale had more statistical cross-informant variance for mother-son dyads than mother-daughter dyads. The Treatment Anxiety subscale on the PedsQL 3.0 Cardiac module includes statements addressing fears about

seeing the doctor, going to the hospital, and having medical treatments (Varni et al., 1999). The results showed that mothers thought their sons were more afraid of the aforementioned elements of treatment than the boys actually were. This could indicate that mothers are less aware of how their sons feel about their treatment, because of a lack of communication about these issues. The fact that the mothers did not know that their sons were not experiencing treatment anxiety may indicate a larger problem of not discussing topics related to their LQTS and related treatments. Mothers also may possibly be influenced by their own anxiety about medical treatment, since their sons are not experiencing this same level of anxiety. It has been found in other research that mothers of children with LQTS have higher levels of anxiety compared to mothers of children with asthma; therefore, the fear and uncertainty of the LQTS diagnosis may have unique implications for mothers of children suffering from this illness (Giuffre et al., 2008).

Even though none of the other subscales were at a statistically significant level in regard to differences between dyads, it noteworthy that mothers of female children believed their daughters had a higher QOL than their daughters' own perceptions on all subscales except one, Cognitive Problems. Conversely, mothers perceived a lower QOL for their sons on all of the subscales except for one, Heart Problems and Treatment.

Hypothesis 2 was not supported at a statistically significant level, since it was underpowered. The findings did show that female children indicated a lower level of QOL overall compared to male children, but not at a statistically significant level. Other studies have found that children with LQTS have more internalizing problems, such as fear of failure and criticism, and that they are less likely to report distressing feelings or share them with peers or family (Giuffre et al., 2008). Therefore, boys with LQTS may

be more likely to underreport difficulties with QOL as a result of the diagnosis, gender-norm expectations, and difficulty with coping emotionally (Calvete et al., 2011). Children with LQTS may be more likely to suppress their fears and anxieties, and may therefore be less likely to report psychosocial stressors.

The data showed that across gender and age, for both children and parents, the lowest mean score was the emotional functioning score. This indicates that both parents and children are reporting that emotional functioning is presently compromised, possibly as a result of LQTS. The emotional functioning scores for this sample were lower than the averages for healthy populations, which implied that those diagnosed with LQTS are at risk for greater emotional distress and diminished quality of life. The emotional functioning subscale measures feelings of fear, anger, sadness, and sleep troubles and worry about the future (Varni et al, 1999).

The data showed that the child age group (8-12) reported lower QOL more often and on more subscales than the young child group (5-7) and the adolescent group (13-19). Considering the developmental age of this group, there may be a connection between developmental transitions such as puberty, social relationship shifts, and school demands that make this a difficult time for children, both emotionally and cognitively. Also, the majority of the individuals in this study were diagnosed before or during this age range; it may therefore be a time when they begin to understand fully the implications of this diagnosis on cognitive and emotional levels. Moreover, the shock of the diagnosis at this age may be more difficult to understand and conceptualize compared to younger children. Another consequence of the LQTS diagnosis for the child is that it sets him or her apart

from other children in their cohort, which may be difficult to manage at an age when homogeneity is socially valued.

### Significance of findings

The findings of the current study, in combination with findings from previous studies, support results indicating that children with LQTS are at greater risk of experiencing more psychosocial stress, and may therefore be more prone to mental health problems (Giuffre et al., 2008).

Due to the unique nature of LQT in regard to its range of symptoms (or lack thereof), it may be important to have a QOL measure that addresses LQTS specifically, and would identify the specific stressors of living with LQTS compared to cardiac problems in general. It is noteworthy from a psychological perspective that LQTS symptomology can range from no symptoms to sudden death, and therefore the unrelenting uncertainty, fear, and vigilance, among other stressors, may play a significant role in degrading QOL (Janney, 2011).

## Limitations

One of the problems with studying LQTS through the lens of chronic illness is that LQTS can be asymptomatic, which could make these findings less generalizable to other chronic illnesses with pain symptoms. Also, fathers were excluded from the present study, which may have interesting implications for cross-informant variance and QOL perceptions. Another limitation of the study is that the measures used were self-report measures, which can be weak in terms of accuracy and consistency. In addition, there was significant variation in the ages of the participants, which could be a confound for this study, because such a broad age sample may not account for other challenges at

various developmental stages. One of the greatest limitations of this study was the small sample size. A larger sample would permit additional statistical analyses, which may have produced more robust findings. Hopefully, this research will be repeated with larger sample sizes across the age groups.

**Future Research Directions**

The needs of this population are great and understanding how to better serve those needs would be cost-and time-efficient. These results allow psychologists to provide more specific help in coping with life as a LQTS patient and in preserving or improving QOL that have far-reaching biopsychosocial effects. Future studies should continue to examine gender differences in children and parents of children with chronic illnesses in regard to QOL and coping, so that a more precise understanding of the unique challenges to this population could be addressed in therapy.

There should also be further research into gender differences in coping and QOL in different cultures. People from different cultures may cope differently with the same illness, and it is important for the field of psychology to understand those unique differences as well. This study examined people seeking treatment from the top Children's hospital in the country, who had the resources to travel to the hospital. There are other populations that are unable to receive that quality of healthcare and may be suffering in different ways. Coping and QOL might be compromised for parents who are unable to get adequate healthcare for their children, and in the case of LQTS, if they are unable to get a diagnosis or genetic testing, their children may die unexpectedly, and they may never know why.

**Gender differences in parents**

Research on differences between mothers and fathers coping with chronic childhood illness has been inconclusive (Sloper, 2000). Some studies found that mothers were more at risk for negative mental health outcomes than fathers, but other studies have found no differences, sometimes because these differences were simply unreported (Sloper, 2000). Some research suggests that fathers are less affected than mothers by serious childhood illness. It is not clear whether these are population differences between men and women, or if there are differences in reactions to the illness based on gender (Sloper, 2000). Research suggests that distress predictors of parents of children with cancer, and in particular mothers, were their assessment of illness-related demands, and of their in their own ability to cope with these were related to distress (Sloper, 2000). It was also found that mothers used more self-directed coping, including self-blame, that was associated with higher levels of maternal distress (Sloper, 2000).

It has been found that there are different predictors of distress in fathers than in mothers coping with the chronic illness of their child. There are stronger effects of risk factors regarding their employment situation, and unemployment stressors, which can result in insecurity, have been shown to negatively affect psychological heath (Sloper, 2000). The results of these stressors are connected to a number of factors: previously raised distress levels before the illness, more vulnerability issues for unemployed fathers, and the impact of employment difficulties caused by the illness on current distress level.

Research on fathers has been more limited than mothers of children with chronic illnesses, and further research is indispensible. Further research should also attempt to explore the differences among parents of children with LQTS, and how they might differ from parents of children with other chronic illnesses. Understanding the unique

challenges of LQTS would be of great interest, since it has specific manifestations (cardiac arrests) that may cause greater psychological stress. The present study did not investigate whether children diagnosed with LQTS were symptomatic or not, which is another important area to delineate and explore in future research.

**Implications of findings**

The findings from this study have implications for healthcare professionals, psychologists, and counselors. Knowing that there are differences between genders and parent's perceptions in relation to QOL for this particular population, the findings might generalize to other populations, such as parents of children with cancer, CF, diabetes, etc., and therefore would be of great significance to a vast number of individuals impacted by chronic illness. Even though LQTS can be asymptomatic, the sheer fact being diagnosed with a chronic illness may be a source of emotional stress for children and families. It will be important in examining the results to investigate specific differences between males and females and dyads and to look for patterns among them. It would be helpful to understand how genders differ in coping and QOL, so that theories could be advanced and built upon in order to create more specific and effective therapeutic interventions.

**Conclusion**

LQTS is garnering attention for its implications for QOL and coping in the families affected. It is a severe condition that can cripple parents with uncertainty, anxiety, stress, and despair. As research continues to grow in this area, it is important to discover the unique characteristics of this illness and their potentially devastating effects on caregivers and children. In creasing research and expanding knowledge about this

population and the gender differences within it will improve psychological attention and care for the mothers and fathers, boys and girls, who will forever deal with the implications of an LQTS diagnosis.

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